Brain Surface Ependymoma With Repeated Episodes of Intratumoral Hemorrhage
—Case Report—

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Abstract
A 70-year-old woman presented with a rare brain surface ependymoma with repeated intratumoral hemorrhage. She was admitted with progressive dementia. Two years earlier, a diagnosis of subcortical hematoma in the right frontal lobe had been made following a fall. On admission, magnetic resonance imaging showed a huge right frontal mass lesion with multiple hemorrhagic cysts. She underwent gross total resection. The tumor was located on the surface of the frontal lobe, and was sharply demarcated from the surrounding brain tissue with no attachment to the ventricular wall. The histological features were consistent with an ependymoma forming perivascular pseudorosettes. Immunohistochemistry showed positive staining for glial fibrillary acidic protein. Electron microscopy showed microvilli and zonula adherens. This case demonstrates the natural course of malignant progression of ectopic ependymomas. Ependymoma should be included in the differential diagnosis of tumors associated with repeated subcortical hematomas, even if located on the brain surface and distant from ventricles.

Key words: brain surface, ependymoma, intratumoral hemorrhage

Introduction
Ependymomas usually arise from a ventricular surface; approximately two-thirds are infratentorial and most of them originate within the fourth ventricle. Before the introduction of computed tomography (CT), supratentorial ependymomas without any visible attachment (ectopic ependymoma) were not considered rare. However, magnetic resonance (MR) imaging now allows accurate evaluation of the exact relationship between the tumor and the ventricular wall and has shown that ectopic localization is exceptional. Only a small number of supratentorial ependymomas located in the cerebral parenchyma with no attachment to the ventricular system have been reported. Most of these tumors were difficult to identify before surgery.

We present a patient with an anaplastic ependymoma occupying the surface of right frontal lobe that manifested as repeated intratumoral hemorrhage. The pre- and postoperative clinical course was monitored by CT during the 3 years that preceded tumor progression.

Case Report
A 70-year-old woman lost consciousness and hit her head on the ground in July 1997. She was transferred to a local hospital by ambulance. Emergent CT demonstrated an atypical intracerebral hematoma in the right frontal lobe (Fig. 1A). The diagnosis was cerebral contusion. She was conservatively treated. She fully recovered and the high-density hematoma had almost completely resolved by August 1997 (Fig. 1B). No abnormal calcification was observed. Follow-up CT was performed in June 1998 at her local hospital. Retrospectively, we recognized a small isodense mass lesion in the right frontal lobe (Fig. 1C). CT in April 1999 demonstrated increased density of the small mass lesion (Fig. 1D) suggesting hemorrhage or calcification, but no further examinations were carried out at her local hospital. She presented at that hospital again in September 2000 because of
progressive dementia and was referred to Kagoshima University Hospital.

On admission neurological examination disclosed memory disturbance and dementia. MR imaging demonstrated a huge intracranial mass lesion separated into multiple cavities in the right frontal lobe (Fig. 2). Some of the cysts showed increased intensity and formation of fluid-level on the T1-weighted images, suggesting intratumoral hemorrhage. The solid portion of the tumor was located between the cystic cavities and was heterogeneously enhanced after intravenous administration of gadolinium-diethylenetriaminepenta-acetic acid. Since the tumor was relatively well demarcated, accompanied by intratumoral hemorrhage, and located at the brain surface, our preoperative diagnosis was anaplastic oligodendroglioma or metastatic brain tumor.

Right free bone flap craniotomy was carried out. The yellow-grayish tumor, located on the surface of the frontal lobe, was clearly demarcated from the surrounding brain tissue (Fig. 3). A number of hematomas or xanthochromic cysts at the frontal tip were not covered by the brain parenchyma. Intraoperative findings confirmed the absence of any attachment to the anterior horn of the lateral ventricle. The tumor was grossly totally resected. Histological examination found pleomorphic tumor cells proliferating in an arrangement of perivascular pseudorosettes (Fig. 4 left). There were some mitoses, and the MIB-1 labeling index was 20% (Fig. 4 right). Immunohistochemical study revealed that the tumor cells were positively stained for glial fibrillary acidic protein, S-100 protein, and vimentin (data not shown). Electron microscopy confirmed the presence of microvilli and zonula adherens (Fig. 5). The appearance of the specimen was typical of ependymoma. Based on these findings, the histological diagnosis was anaplastic ependymoma.

Her postoperative course was not favorable. She required a second operation because of postoperative bleeding into the cavity as a result of removal of the tumor. Although she recovered and is free of
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Fig. 3 Intraoperative photograph demonstrating the cystic tumor located on the surface of the frontal lobe, and clearly demarcated from the surrounding brain tissue.

Fig. 4 Photomicrographs showing pleomorphic tumor cells proliferating in an arrangement of perivascular pseudorosettes (left: HE stain, original magnification ×100), and the MIB-1 labeling index was 20% (right: original magnification ×100).

Fig. 5 Electron micrograph revealing tumor cells with zonula adherens (arrowheads) and a cluster of microvilli (arrow). Bar = 1 μm.

Discussion

The present patient with a rare brain surface ependymoma had experienced at least three episodes of intratumoral hemorrhage. Due to exophytic growth, the tumor was not covered by the brain parenchyma. Brain surface ependymomas are very rare, with only three cases reported previously (Table 1).3,7,8)

<table>
<thead>
<tr>
<th>Author (Year)</th>
<th>Age/Sex</th>
<th>Tumor location</th>
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<tbody>
<tr>
<td>Vernet et al. (1995)</td>
<td>11/F</td>
<td>Lt frontal lobe</td>
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<tr>
<td>Fujimoto et al. (1999)</td>
<td>13/M</td>
<td>Lt parietal lobe</td>
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<tr>
<td>Sato et al. (2000)</td>
<td>41/F</td>
<td>Lt frontoparietal lobe</td>
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<tr>
<td>Present case</td>
<td>70/F</td>
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The pathogenesis of brain surface ependymomas remains unclear. Currently, there are three tumor recurrence as of November 2001, she remains bedridden.

Table 1 Reported cases of brain surface ependymoma

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hypotheses for the ectopic origin of ependymomas.8) The tumor could develop from an intraparenchymal or subarachnoid ependymal cyst; from a ventricular pouch; or from a heterotopic ependymal nest resulting from disorders in migration from the germinal matrix. In our case, the ventricular contour was intact so that the second mechanism seems improbable.

None of the previously reported brain surface ependymomas were associated with intratumoral hemorrhage, which made the differential diagnosis even more difficult in our case. Intratumoral hemorrhage in supratentorial ependymoma is a rare event, although spinal subarachnoid hemorrhage is seen in some patients with a variant form, myxopapillary ependymoma of the cauda equina. Interestingly, our patient had experienced at least three episodes of intracerebral hemorrhage during the 2 years that preceded her admission to our institution. These hemorrhages occurred in the right frontal lobe where the large cystic tumor was located. The initial hemorrhage in July 1997 (Fig. 1A) was treated as a contusional hemorrhage because it was apparently the result of a head injury when she fell after losing consciousness. However, our retrospective study of the CT scans taken in July 1997 showed that the mass effect of the hematoma was smaller than would be expected, suggesting that the hematoma was not associated with contusion of the surrounding tissue. In addition, the follow-up CT scan obtained in June 1998 (Fig. 1C) showed an isodense mass lesion in the same location as the previous hematoma. This small mass showed increased density suggesting repeated hemorrhage. Therefore, we conclude that the small tumor had developed on the brain surface and rapidly grew during the year preceding the operation. At surgical removal in April 1999, the MIB-1 labeling index was 20% with many mitoses, suggesting that malignant transformation had occurred during the follow-up period.

This case indicates that ependymoma should be considered in the differential diagnosis of supratentorial tumor with intratumoral hemorrhage, even if not in contact with the ventricular system. Since the extent of surgical removal is a good prognostic factor in patients with ependymomas, intraoperative histological diagnosis with frozen sections is exceedingly useful for helping to determine the advisability of aggressive surgical removal.

References


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